

The Genetics of Hearing Loss in the Palestinian Population

Why Hearing Loss?

Hearing Loss in the Palestinian Population*

Gender	Population	Affected Individuals	Percentage
Females	1,495,055	2,410	0.16
Males	1,524,649	2,885	0.18

*Does not include those parts of Jerusalem annexed by Israel in 1967.
Palestinian Central Bureau of Statistics, 1999. Population, Housing and Establishment Census 1997, Statistical Brief. (Summary of Census Results) Ramallah -Palestine.

Hearing loss affecting the Palestinians is severe to profound bilateral congenital sensorineural in nature

Sample audiogram of an affected individual from Family K

Classification of Hearing Loss

Hereditary Research Laboratory

- There is no policy yet that addresses hearing loss as a public health threat.
 - Limited number of specialized schools for the deaf
 - Education is limited to up to the age of 16
 - Chances for rehabilitation are minimum and limited to vocational training
 - Lack of database
- The HRL is interested in the basic science that addresses relevant problems and ties it to the new advancement in the field of genetics

The Inner Ear

Hypothetical uncoiling of cochlea

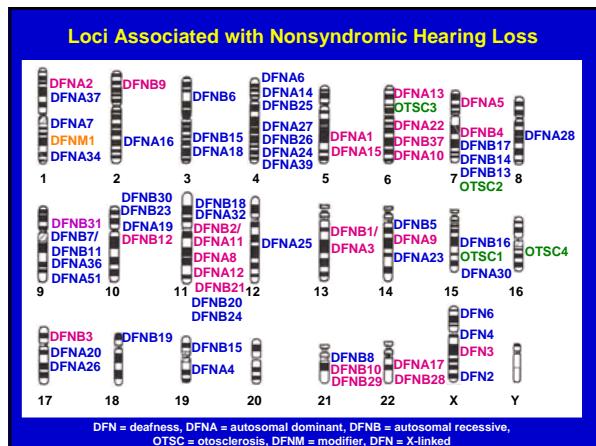
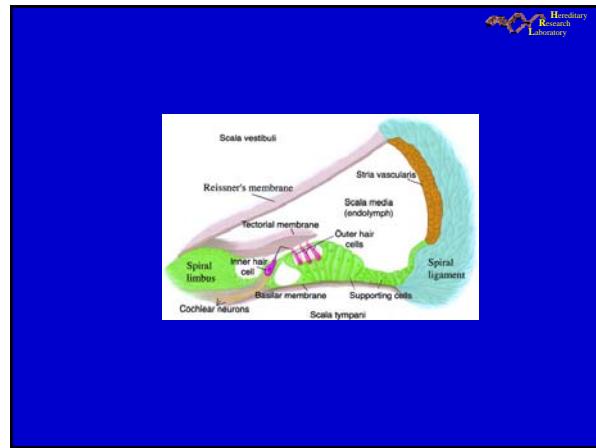
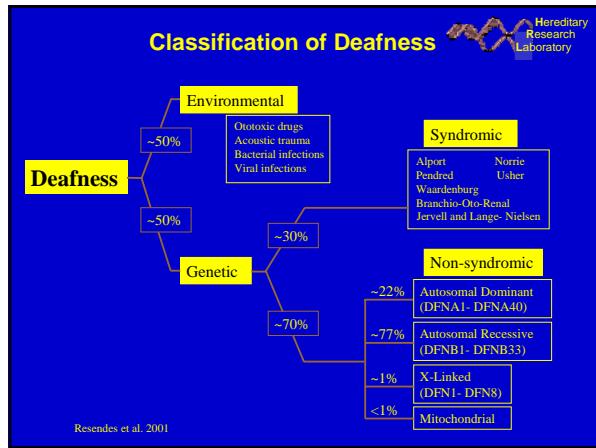
Cross section of cochlea

Top view of hair cell, showing stereocilia projecting from top of hair cell.

Hereditary Research Laboratory

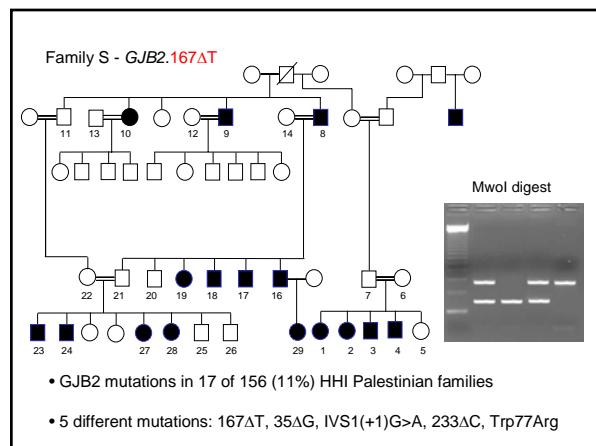
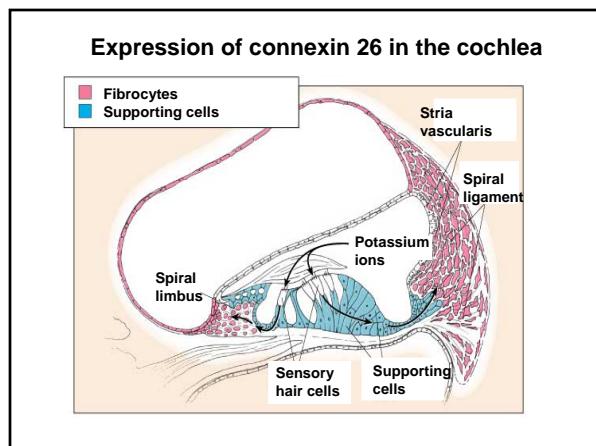
Hair Cells – Function

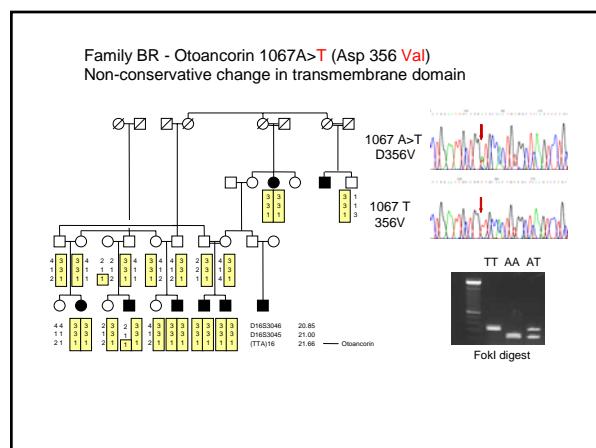
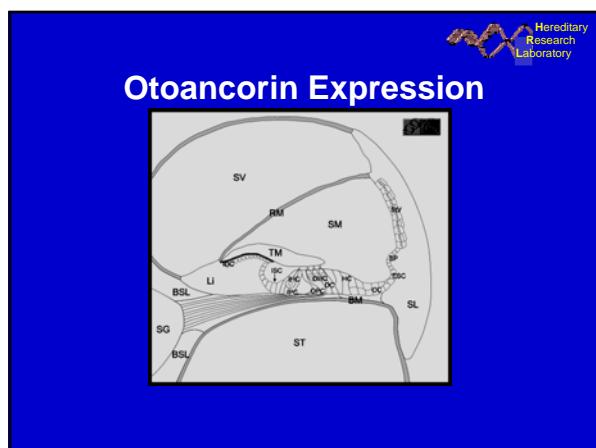
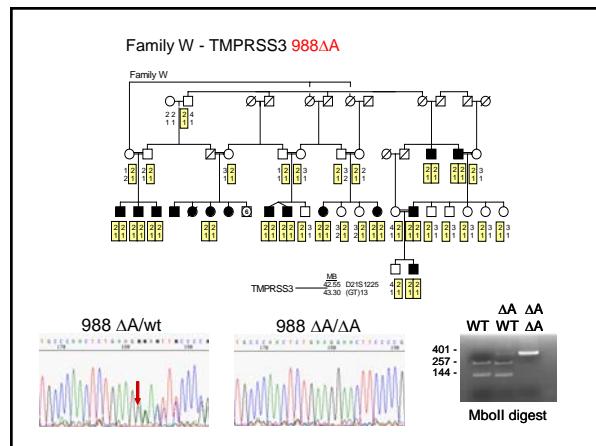
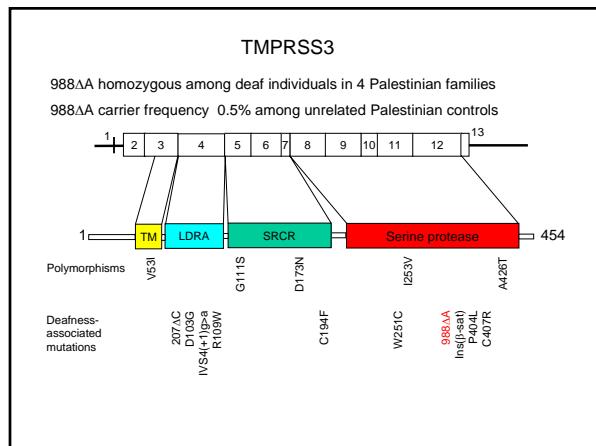
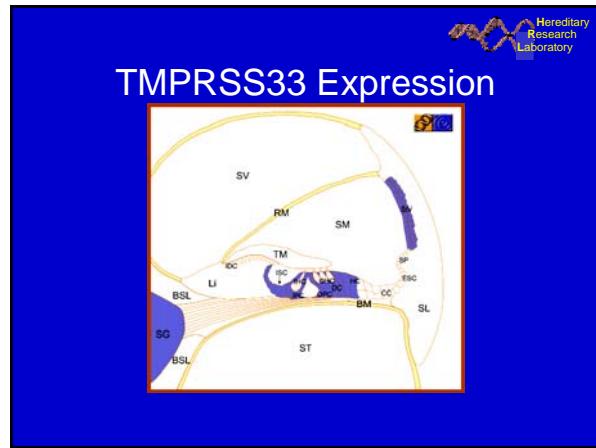
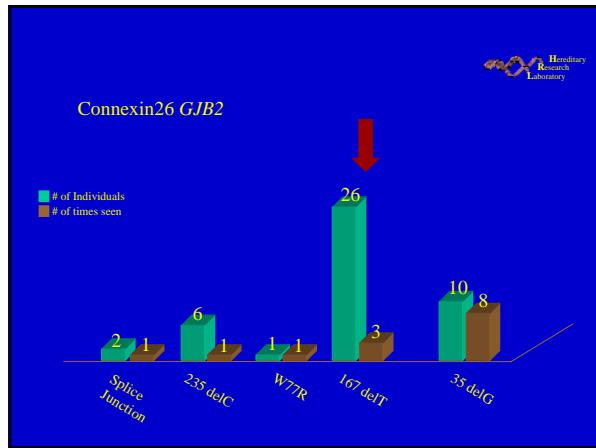
Auditory/Vestibular stimulus → Stereocilia deflection → Opening of ion channels & entrance of Ca++ and K+ ions → Hair cell depolarization → Neurotransmitter release at synapse

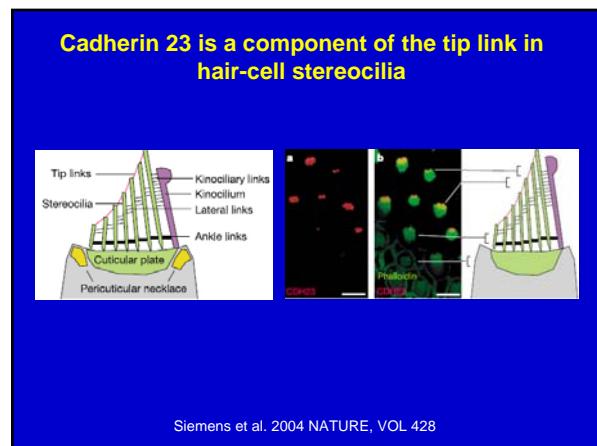
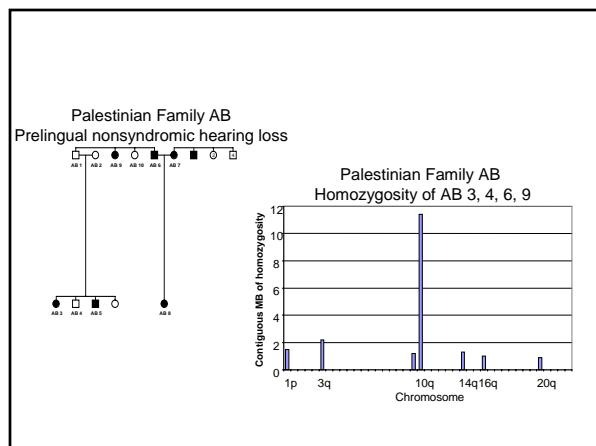
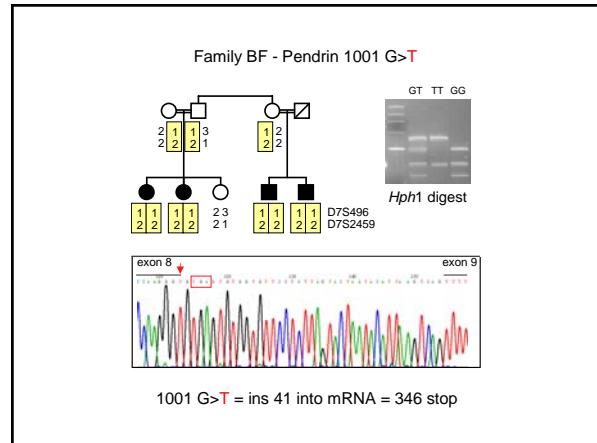
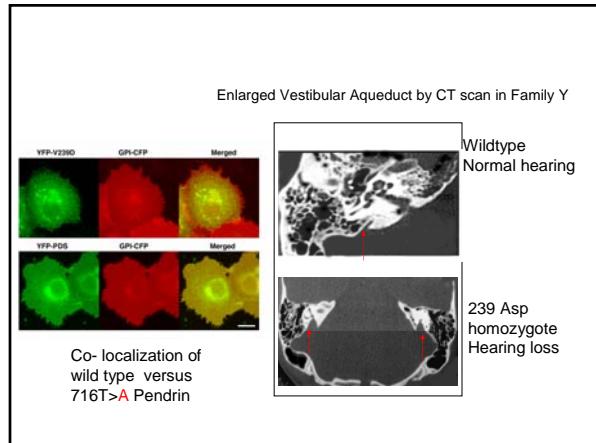
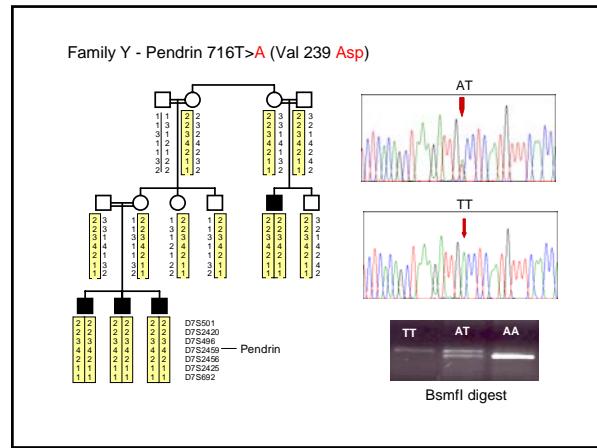
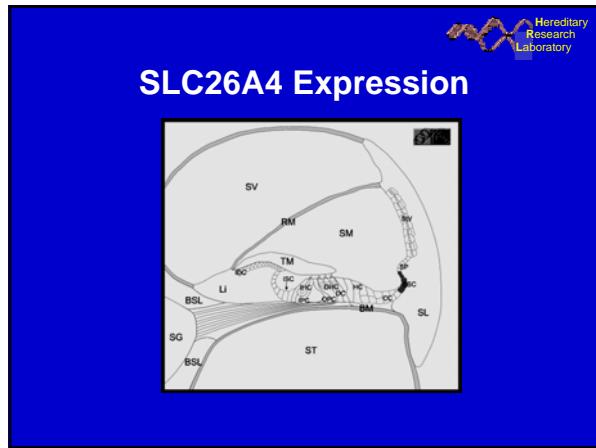


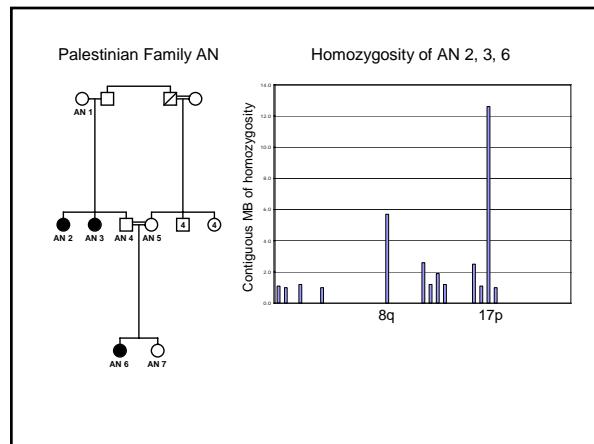
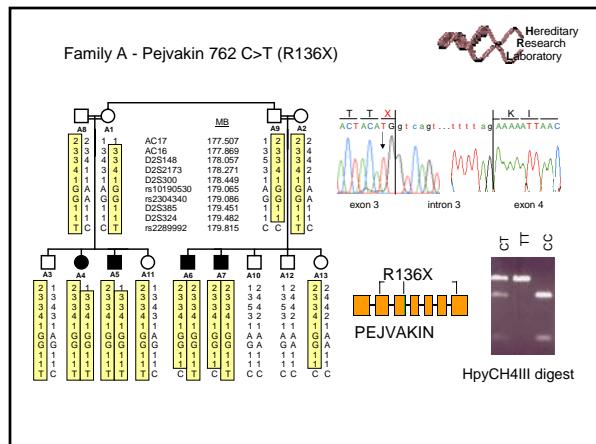
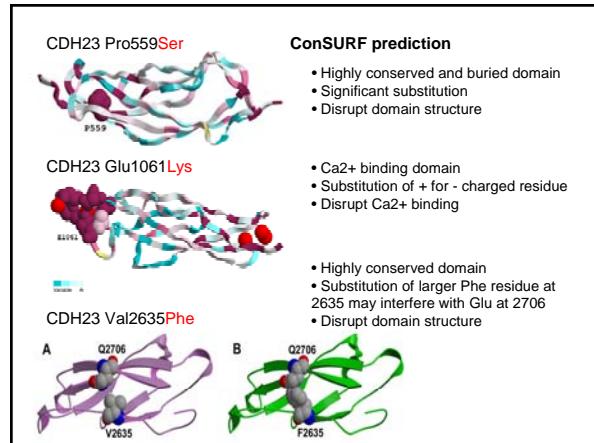
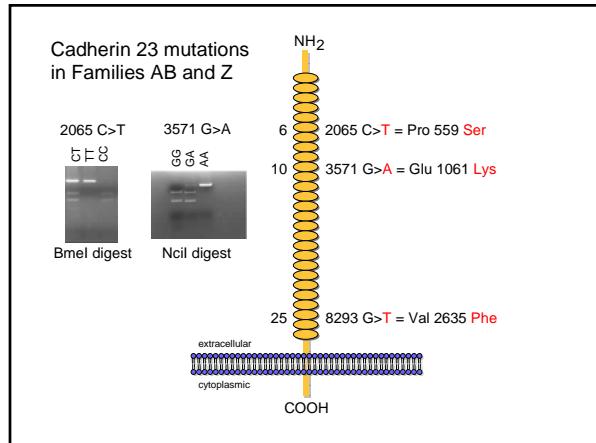
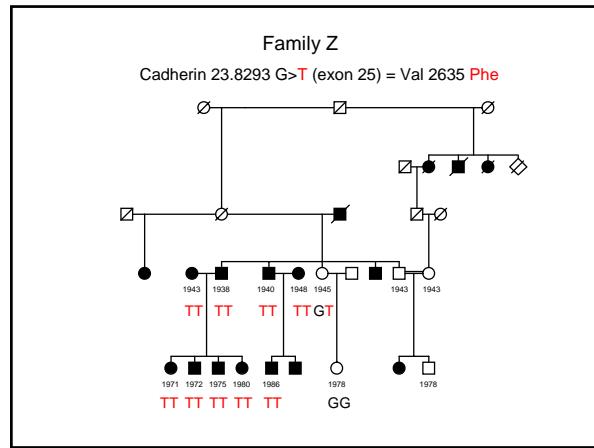
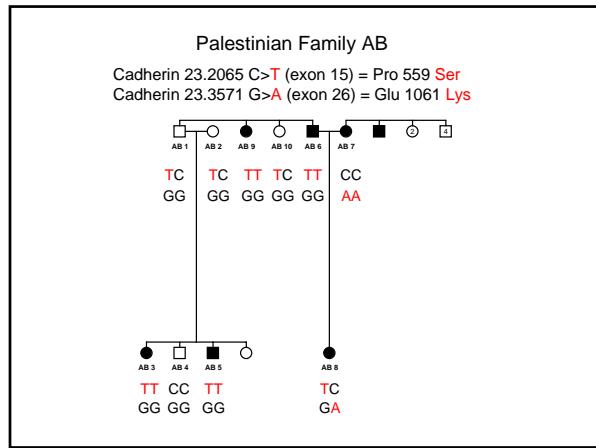
Genes for Hearing Loss in Middle Eastern Families in our Project

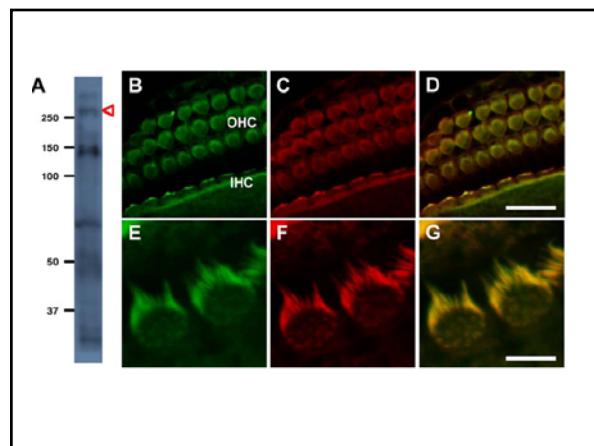
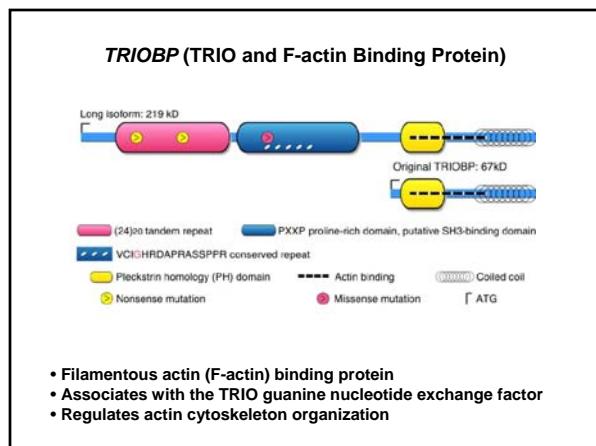
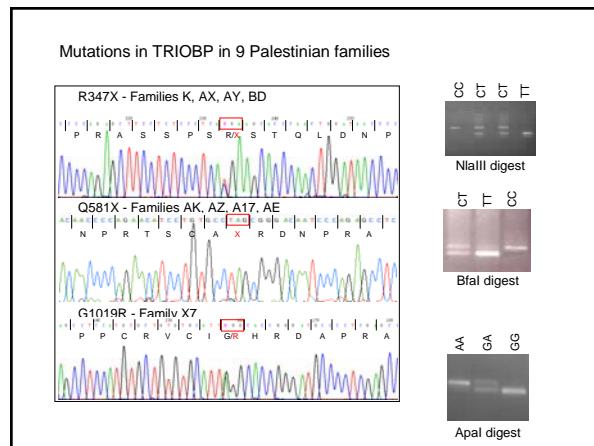
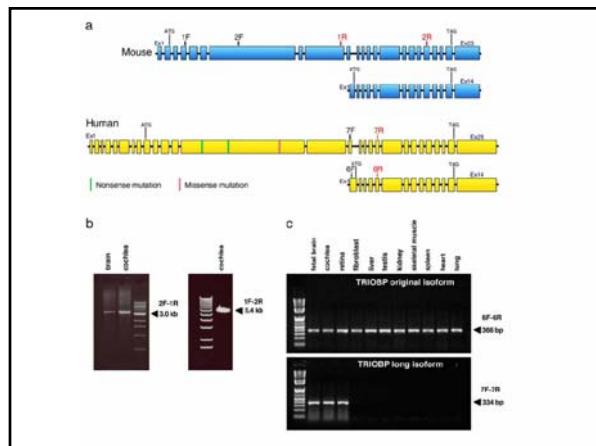
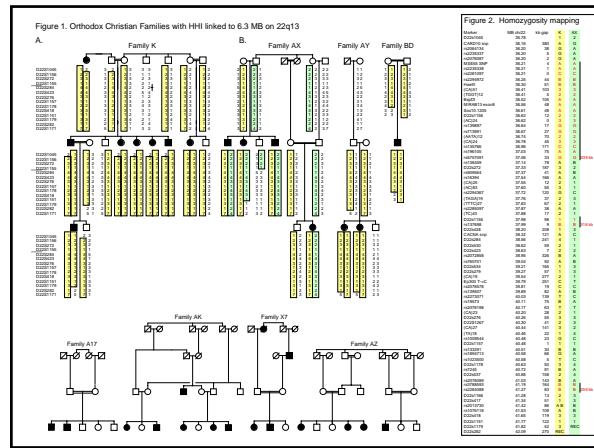
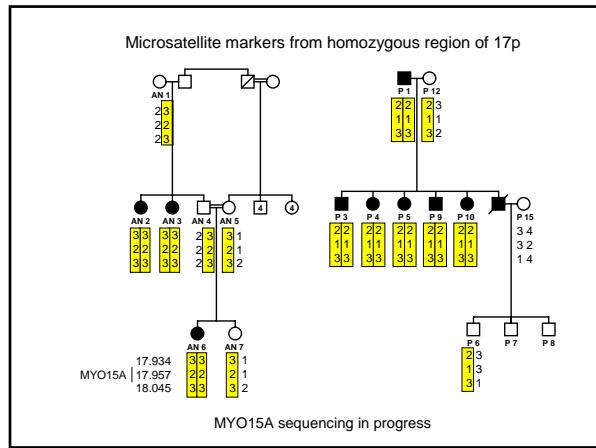
Connexin 26	Pendrin
★ POU4F3	Cadherin 23
★ MYO3A	MYO15A
TMPRSS3	★ TRIOBP
Otoancorin	Pejvakin
Others still unknown	

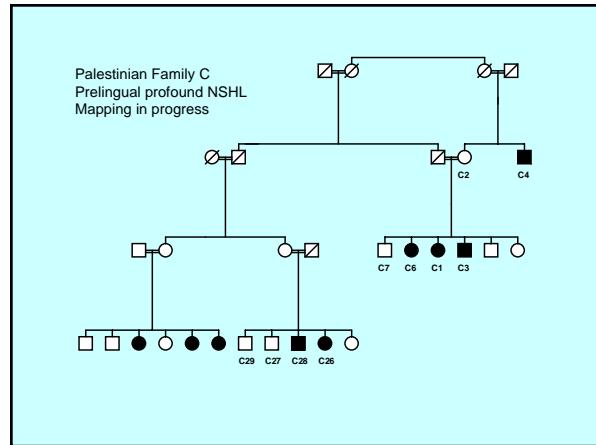
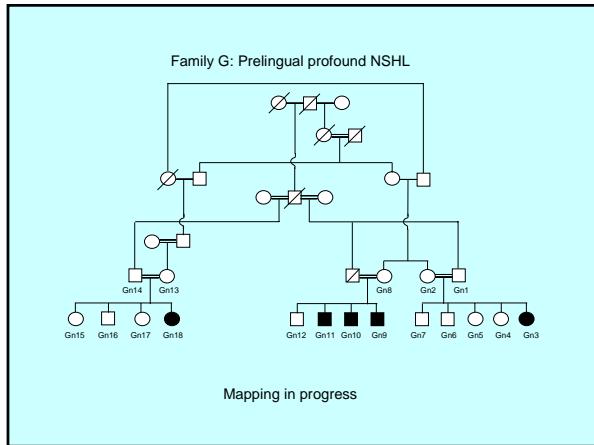
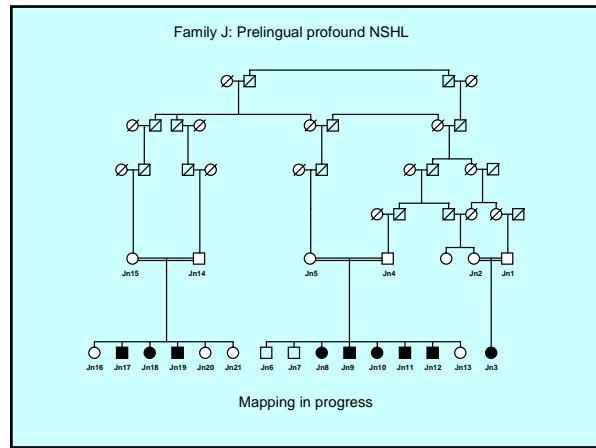
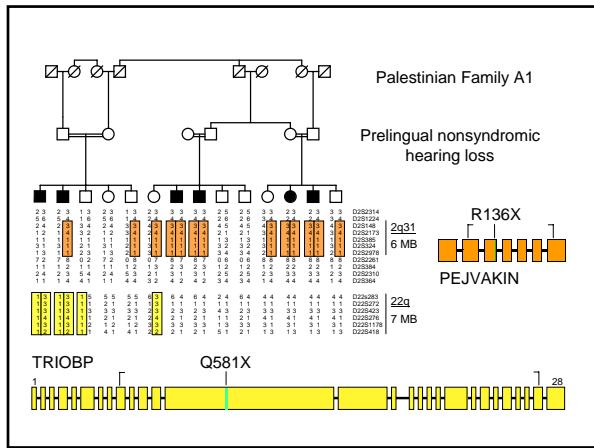












Conclusions



- Inherited hearing loss in the Palestinian population is highly heterogeneous, both in the number of genes involved and in the number of alleles at each gene.
- Alleles responsible for hearing loss in this population are individually very rare.
- Families with hearing loss who are wildtype for *GJB2* are likely to harbor novel alleles, either of known hearing-related genes (as in these three families) or of previously unknown genes.
- We have identified and cloned a new gene (TRIOBP) that underlines hearing loss in nine Palestinian families.
- Studies of multiple Palestinian kindreds are in progress to identify and characterize yet unknown genes.

Genetics of Hearing Loss in Middle Eastern Families



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Supported by NIH RO1 DC005641